LV in the subacute phase(1–6 months after the onset of myocarditis), and inflammatory cells(LCA cells) identified with immunoperoxidase staining using anti-leukocyte common antigen serum were counted in high power field of all samples. The LV end diastolic dimension(LVDd) and ejection fraction(EF) were measured by the echocardiography at the biopsy(B) and one year later(F). There was a significant correlation between the mean number of LCA positive cells and the %change of LVDd ($r = 0.551$, $p < 0.05$). 16 pts were classified by the average number of LCA cells into Group-I($n = 7$) with 1.0 or more(/HPF) LCA cells and Group-II($n = 9$) with LCA cells less than 1.0/HPF. The results were as shown below.

	LCAcells(/HPF)	B-LVDd(mm)/EF	F-LVDd(mm)/EF
Group-I	2.6 ± 0.8	57.5 ± 5.1/0.53 ± 0.09	57.6 ± 5.6/0.51 ± 0.12
Group-II	0.6 ± 0.2	55.1 ± 5.3/0.51 ± 0.08	50.8 ± 6.4*/0.59 ± 0.08

*$p < 0.05$ vs Group-I

There was no significant difference in the period from the onset of myocarditis to the biopsy (Group-I vs Group-II; 3.0 ± 1.9 vs 1.9 ± 1.6 months). LVDd decreased in Group-II 1 year after biopsy more than in Group-I. Thus, these results indicate that the improvement of cardiac function in acute myocarditis might be predicted by the degree of residual LCA infiltration in subacute phase(over one month after the onset).

1033-71 Circulating Cardiac Autoantibodies as Autoimmune Markers in Clinical and Biopsy-Proven Myocarditis

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Myocarditis and dilated cardiomyopathy (DCM) may be phases of an ongoing autoimmune disease of the myocardium. Cardiac autoantibodies (Abs) are found in 30–40% of DCM patients. Their detection in myocarditis would provide evidence for autoimmunity. Cardiac antibody status was assessed in 53 patients from the Myocarditis Treatment Trial (35 male, aged 42 ± 15 years). The antibody specificities included organ-specific and skeletal muscle cross-reactive Abs by indirect immunofluorescence and anti-α myosin Abs by enzyme-linked immunosorbent assay (ELISA). All patients had clinical myocarditis and unexplained heart failure, but only 24 were classified as histological myocarditis (Dallas criteria) on endomyocardial biopsy and randomised in the Trial. By immunofluorescence cardiac Abs were more common in myocarditis patients than in normals (13/53, 24% vs 18/200, 9%, $p = 0.004$); by ELISA abnormally raised anti-α myosin Abs were also more frequent than in normals (9/53, 17% vs 1/52, 2%, $p = 0.009$); 18 patients (34%) had a positive result with one or both tests. Similar proportions of patients with and without histological myocarditis contained Abs by immunofluorescence (8/24 vs 5/29 respectively, p = NS) and by ELISA (4/24 vs 5/29 respectively, p = NS). Cardiac Abs are found in 34% of patients with clinical myocarditis; this provides evidence for autoimmune involvement. The lack of correlation of antibody status with the histological diagnosis of myocarditis suggests that there may be inaccuracy when diagnosis is made on histology alone. Autoimmune markers may provide adjunct diagnostic tools and identify myocarditis patients in whom immunosuppression is of potential benefit.

1033-72 Preliminary Report of the Multicenter Giant Cell Myocarditis Study Group

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Idiopathic Giant Cell Myocarditis is a rare and frequently fatal disease of unknown cause which has only been reported in isolated cases and two small series. Accordingly, research on this disease is hampered by a lack of data. In January 1995, we established a multicenter study group to better address the many unresolved questions about the natural history and treatment of idiopathic giant cell myocarditis. Direct mailing to cardiovascular centers worldwide and journal announcements produced 42 cases ranging in age from 39 days to 70 years. Most present with congestive heart failure or ventricular dysrhythmias. Without immunosuppression the clinical course is one of rapid deterioration to death or heart transplant. Several patients survived longterm (range 3–9 yrs) after immunosuppressive treatment. Thirteen patients in our registry underwent heart transplant. Eleven are alive up to 12 years post-transplant, 2 with histologic disease recurrence. This is by far the largest cohort of this rare disease ever reported and represents the collective experience of many medical centers worldwide. We hope to present a final report at future scientific sessions.

1033-73 Arrhythmogenic Right Ventricular Dysplasia-Cardiomyopathy: A Form of Healing Myocarditis?

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To determine the role of inflammation and fibrosis in right ventricular dysplasia (RVD), we histologically evaluated 15 hearts from patients with RVD dying suddenly, and 11 age-matched control hearts. RVD was defined as right ventricular dilatation with focal fibrosis or thinning < 0.5 mm. Eight histologic sections were taken from each ventricle, apex to base, stained for collagen, and the degree of fat infiltration, fibrosis, and number of inflammatory foci quantitated. The mean age of RVD was 31 ± 10 years. 4 cases (27%) were familial, 6 patients had a history of arrhythmias (40%), and 8 deaths occurred during exercise (53%). Foci of inflammation were present in 14/15 cases (93%)(mean number 15 ± 5.2), and microscopic foci of left ventricular fibrosis and/or inflammation were found in 12 cases (80%). Inflammation correlated negatively with age of the patient ($p = 0.02$, r^2 0.4), and fibrosis showed a positive correlation with age ($p = 0.008$, r^2 0.5). Left ventricular fibrosis was greatest in the base of the free wall (12 ± 3%) and least in the ventricular septum (6.6 ± 0.9%, $p = 0.03$). Fibrosis was diffuse in the right ventricle (mean 20 ± 5%) without predilection for site. No differences in the degree of fibrosis or inflammation were noted with respect to family history, exercise at death, or history of arrhythmias. Compared to controls, RVD was hearts had greater fibrosis ($p = 0.001$) and inflammation ($p = 0.0001$), but there was no significant difference in degree of fat infiltration of the right ventricle (34 ± 4.8% RVD vs. 23 ± 5.6% control, $p = 0.1$). RVD is an inflammatory disease that progresses to fibrosis and often involves both ventricles; fat infiltration is a secondary phenomenon.

1033-74 Arrhythmogenic Right Ventricular Cardiomyopathy: Dysplasia, Dystrophy or Myocarditis?

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Aim of the investigation was to elucidate the nature of the pathobiological events underlying arrhythmogenic right ventricular cardiomyopathy (ARVC). 30 hearts with ARVC were studied (20 M, 10 F, aged 15–65 years, mean 28). Source of specimens was autopsy in 27 and cardiac transplantation in 3. Mode of death of autopsy cases was sudden in 24 and congestive heart failure in 3 (due to cerebral thromboembolism in 1). Previous symptoms in terms of syncope, palpitations or heart failure, were complained by 17 patients (57%). Basal ECG, available in 19, showed inverted T-waves in the right precordial leads in 15 (79%) and ventricular arrhythmias in 15 (79%). RV aneurysms were present in 15 hearts (50%), mostly located in the inferior wall. Involvement of the left ventricle was observed in 14 cases (47%) and in 6 of them was extended to the ventricular septum. Scattered foci of T-cell lymphocytes with myocyte necrosis were present in 20 cases (67%). On the basis of the histopathologic substrate, we observed a fatty (40%) and a fibrous-fatty (60%) patterns. The fibrous-fatty pattern showed a higher incidence of ventricular arrhythmias ($p = 0.05$), a thinner RV wall ($p < 0.0001$), and a higher occurrence of focal myocarditis, left ventricular involvement and RV aneurysms ($p < 0.001$). In conclusion, myocardial atrophy observed in the fibrous-fatty variety of ARVC, appears to be the consequence of an acquired injury (necrosis) and repair (fibrous-fatty replacement) progressive process, mediated by patchy myocarditis; whether inflammation is a primary event or reactive to spontaneous necrosis remains obscure. A programmed cell death or apoptosis in the setting of postnatal involution of the RV might be considered.

1033-75 Chagas' Heart Patients Without Cardiac Enlargement Have Impaired Epicardial Coronary Vasodilation but No Vasotonic Angina

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Myocardial perfusion defects are detected in many Chagas' heart patients (CHP) and coronary spasm has been postulated to cause these disturbances. In pts with vasotonic angina (VA), hyperventilation (H) provokes vasoconstriction responsive to nitrates. Aim of this study was to measure vasomotor epicardial coronary responses to H and isosorbide dinitrate (ISDN), in 24 CHP (age 56 ± 9, 14 M), 15 with (D-CHP) and 9 without cardiac dilation (ND-CHP), and 9 VA pts with angiographycally normal coronary arteries (age 45 ± 9, 3 M). Quantitative angiography was performed at baseline (B), after H, and after administration of 5 mg of ISDN, in proximal (LADP) and distal (LADD) segments of the left anterior descending coronary artery and proximal segments of left circunflex artery (CXP). No patients had clinical or EKG ischemic changes during H, and never was local or diffuse spasm detected.